EPI-NEWS NATIONAL SURVEILLANCE OF COMMUNICABLE DISEASES
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RISK OF VARIANT CREUTZFELDT-JAKOB DISEASE IN DENMARK

No. 12, 2001

Classical Creutzfeldt-Jakob disease (CJD) was described in the 1920's, while the first human case of variant CJD (vCJD) was reported in the UK in 1995. Since then the world total of vCJD cases amounts to 99, 95 diagnosed in the UK, three in France and one in Ireland. No case of vCJD has been detected in Denmark, EPI-NEWS 48/2000. vCJD is attributed to food-borne infection from cattle with mad cow disease (bovine spongiform encephalopathy or BSE).

Prion diseases in man

CJD belongs to the group of prion diseases and may be sporadic, familial or infectious. The sporadic form, which makes up about 85% of all CJD cases, has an annual incidence of about 1 per million population throughout the world. Sporadic CJD is thought to arise when the normally occurring prion protein in brain nerve cells undergoes a spontaneous conformational change to adopt a form that is resistant to enzymatic breakdown. The altered prion protein also has the capacity to induce the same conformational change in normal prion proteins by an autocatalytic process. This leads to an accumulation of altered prion proteins in the nerve cells, which degenerate and die, producing small holes in the cerebral grey matter. The resulting spongy appearance gives rise to the term "spongiform encephalopathy". Familial CJD is caused by destabilizing mutations in the gene encoding the prion protein. Infectious CJD is due to the administration or ingestion of the disease-producing prions, which initiate the autocatalytic process. The group of infectious prion diseases includes CJD from the use of transplants or medicines derived from cadavers with undiagnosed CJD, kuru and vCJD.

The British vCJD epidemic

The number of cases of vCJD in the UK is rising. However, it is still not possible to estimate the potential extent of the epidemic. From various estimates of factors such as incubation period and number of exposed individuals, it can be calculated that a total of somewhere between 500 and 200,000 cases could occur. The model assumes that transmission ceased after 1990. There can still be many cases in the period of incubation, which is estimated to last from a few years to over 40 years. The incubation period for infectious prion diseases is known to depend on the infective dose, the route of infection and the genetic predisposition of the recipient.

Diagnosis of vCJD

vCJD declares itself by progressive neuropsychiatric symptoms, often accompanied by cerebellar signs with ataxia and myoclonus. Later a progressive dementia sets in. Patients with vCJD are much younger (mean age about 29 years) than patients with classical CJD (mean age about 65 years). However, vCJD has recently been diagnosed in a man aged over 70 years. The definitive diagnosis of vCJD, as that of classical CJD, can only be made at autopsy, by demonstrating the characteristic changes in the brain. There are no clinical tests that can firmly diagnose vCJD while the patient is alive. Brain biopsy is not recommended. The detection of various neuron-specific proteins in cerebrospinal fluid can support a suspected diagnosis, as can EEG and MR scanning. Further details were given in EPI-NEWS 9/00. It has been reported that disease-producing prions can be demonstrated in tonsillar biopsies from pa-tients with vCJD, but the method has not yet been fully validated. It should be stressed that tonsillar biopsy cannot be used to diagnose classical CJD.

Risk of infection with vCJD

Prions are very resistant to standard physical and chemical inactivation methods. Thus prions are not neutralized by normal autoclaving for 15 min. at 121°C. In Britain it is recommended that instruments used in invasive procedures in patients with suspected vCJD should be discarded. There is no evidence to suggest that care staff are at increased risk of occupational infection with either classical CJD or vCJD, and the diseases are not transmitted by normal social contact, including sexual intercourse. A working group is currently revising the Danish National Board of Health guidelines on hygienic precautions in CJD. vCJD is now regarded as a primarily food-borne disease. By far the greatest infective risk is found in the so-called specified risk materials (SRM) from infected animals. SRM comprise the skull, brain, spinal cord, eyes, tonsils and intestines. It is impossible to calculate with any certainty how many Danes have been exposed to infection from foods containing SRM before these were banned. Many Danes have been to the UK during the 1980's and may have been exposed to infection there, and on that basis alone it is impossible to discount the occurrence of vCJD cases in Denmark. There is also a risk of infection from other sources, at least in theory. The infective prion is found in lymphoid tissue and white blood cells. Animal experiments have shown that BSE can be transmitted by blood transfusion, which it was not thought that classical CJD could be. Some countries have excluded from donating blood any person who has spent more than 6 months in Britain during the period 1980-1996. The preparation of plasma products from British blood has been stopped in the UK and leukocyte depletion of blood preparations has been introduced. Leukocyte depletion has also been introduced in other countries, although the relative risk of transmitting prions via blood and the effect of leukocyte depletion are poorly understood. In Denmark the authorities are considering actions to be taken regarding blood.

Prevention of vCJD

As vCJD is primarily a food-borne disease, the most important preventive measure is to ensure that foods are BSE-free. This is undertaken by avoiding the creation of further BSE-infected cattle, i.e. by prohibiting the use of meat and bone meal in fodder. A further step is to try to prevent the use of BSE-infected cattle as human food, which should be aided by screening all slaughtered cattle over 30 months of age for prions. Finally, banning the use of SRM in food production safeguards against using the potentially most infectious material for human consumption. Prion diseases in cattle will be presented in a future issue of EPI-NEWS.

(E. Smith, N. Strandberg Pedersen, Department of Epidemiology)

21 March 2001

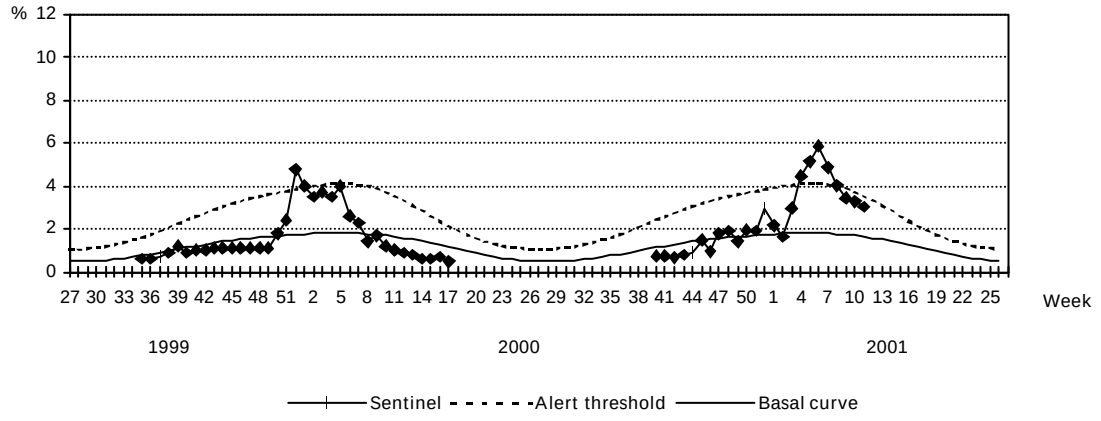
Patients with positive cultures of pathogenic intestinal bacteria in 2001, by county

	Campylobacter		Yersinia enteritidis		S. typhimurium		S. enteritidis		Other zoon. Salmonella spp.	
	Jan	Feb	Jan	Feb	Jan	Feb	Jan	Feb	Jan	Feb
Cph. Municip.	29	32	1	-	4	2	3	1	7	8
Frb. Municip.	5	1	-	-	-	-	-	-	-	-
Copenhagen	28	21	2	-	4	2	3	2	7	7
Frederiksborg	13	10	-	1	2	1	6	5	4	4
Roskilde	10	8	1	-	2	1	2	1	2	4
West Zealand	5	3	-	2	2	1	3	4	3	1
Storstrøms	10	8	3	-	5	1	1	1	4	1
Bornholms	1	1	-	-	-	-	-	-	-	-
Funen	17	13	3	-	7	2	6	2	3	6
South Jutland	11	2	-	-	2	1	3	8	-	2
Ribe	9	14	1	-	-	-	5	3	5	1
Vejle	13	14	2	1	2	1	8	11	1	4
Ringkøbing	17	15	2	1	-	1	3	1	-	2
Aarhus	27	14	1	2	3	2	6	8	4	2
Viborg	3	5	-	-	1	1	5	3	1	1
North Jutland	15	11	3	1	1	1	4	1	2	5
Unknown	-	-	-	-	-	-	1	-	-	-
DK Jan/Feb 2001	213	172	19	8	35	17	59	51	43	48
DK Jan/Feb 2000	176	187	23	8	14	23	67	53	41	41

(Intestinal Bacteriology Lab.)

Sentinel surveillance of influenza activity

Weekly percentage of consultations, 1999/2000/2001



Sentinel: Influenza consultations as % of total consultations

Basal curve: Expected frequency of influenza consultations under non-epidemic conditions

Alert threshold: Possible incipient epidemic

(Dept. of Epidemiology)